Pre-pubertal Presentation of Peritoneal Inclusion Cyst Associated with Congenital Lower Extremity Venous Valve Agenesis

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ABSTRACT

Peritoneal inclusion cysts are uncommon lesions that usually occur in the pelvis of reproductive-age females. The case of a 7-year-old girl with an inflamed peritoneal inclusion cyst with unusual right paracolic localization and congenital lower extremity superficial and deep venous valve agenesis is presented. Inflammation of the peritoneal inclusion cyst was responsible for the signs of acute abdomen and subsequent presentation at our center. The cystic structure was initially diagnosed using ultrasonography, and its complete extent (8cm x 6.5cm x 4cm) was evident after magnetic resonance imaging. The minimal access approach was opted for to resect the entire cyst from the lateral border of the ascending colon. Afterwards, the cyst was punctured to reduce its size and to retrieve the cyst wall using an endoscopic specimen retrieval bag. Minimal access surgery precautions in this patient with congenital lower extremity venous valve agenesis are discussed.

Key Words: Peritoneal inclusion cyst, Prepubertal females, Diagnosis, Laparoscopy, Venous valve agenesis.

INTRODUCTION

Peritoneal inclusion cysts are uncommon lesions that occur in the pelvis of reproductive-age females with the average age of 32 years (range, 17 to 61) at the time of diagnosis.[1,2] Furthermore, in adult females a prior history of abdominal surgery, pelvic inflammatory disease, or endometriosis is present. The common presenting symptom is lower abdominal or pelvic pain. Acute presentations, frequently simulating appendicitis, have been described in adult females diagnosed with peritoneal inclusion cysts in previous reports.[3,4] The cysts are often multiple and can range from 1cm to 20cm in diameter with some forming confluent masses that cover the entire pelvis. Peritoneal inclusion cysts can be free floating in the abdomen or adherent to pelvic or abdominal structures. There are no reports in the literature to date on peritoneal inclusion cysts in a prepubertal female. Also, the literature is void on the minimal access management of patients with congenital lower extremity venous valve agenesis.

CASE REPORT

A 7-year-old girl was referred with acute, severe, right lower quadrant abdominal pain with symptoms similar to those of acute appendicitis. The girl had a history of congenital agenesis of the superficial and deep venous valves of the lower extremities. An ultrasound examination was performed that detected a large cystic structure in the right paracolic recess. Further investigations with magnetic resonance imaging revealed the full size (8cm x 6.5cm x 4cm) and extension (lateral paracolic extension from the cecum to the hepatic flexure along the length of the ascending colon) of the cyst **(Figure 1)**. The laboratory investigations were not remarkable, and infection parameters were elevated. Tumor markers were normal: α-feto protein (AFP)-1.4ng/mL (normal <15), carcino embryonic antigen (CEA)-1.3ng/mL (normal <5), and ß-Human chorionic gonadotropin (ßhCG)-2mIU/mL (normal <50).

With the differential diagnosis of colonic duplication, mesenterial cyst, or peritoneal inclusion cyst, a minimal access approach was opted to diagnose and resect the cystic structure. The patient was placed in a supine

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position, and the procedure was performed using a 5-mm 30° scope along with two 5-mm instruments. Access was gained into the abdominal cavity by using an open access technique with an incision through the left umbilical fold and the open placement of a 10-mm port for the scope. Insufflation pressure was maintained at 8mm Hg with a flow of 0.5L/m considering the congenital lower extremity valve agenesis manifestation, to prevent excessive venous stasis of the lower extremities during the procedure. Two 5-mm working ports were introduced into the upper abdominal and lower abdominal cavity under vision.

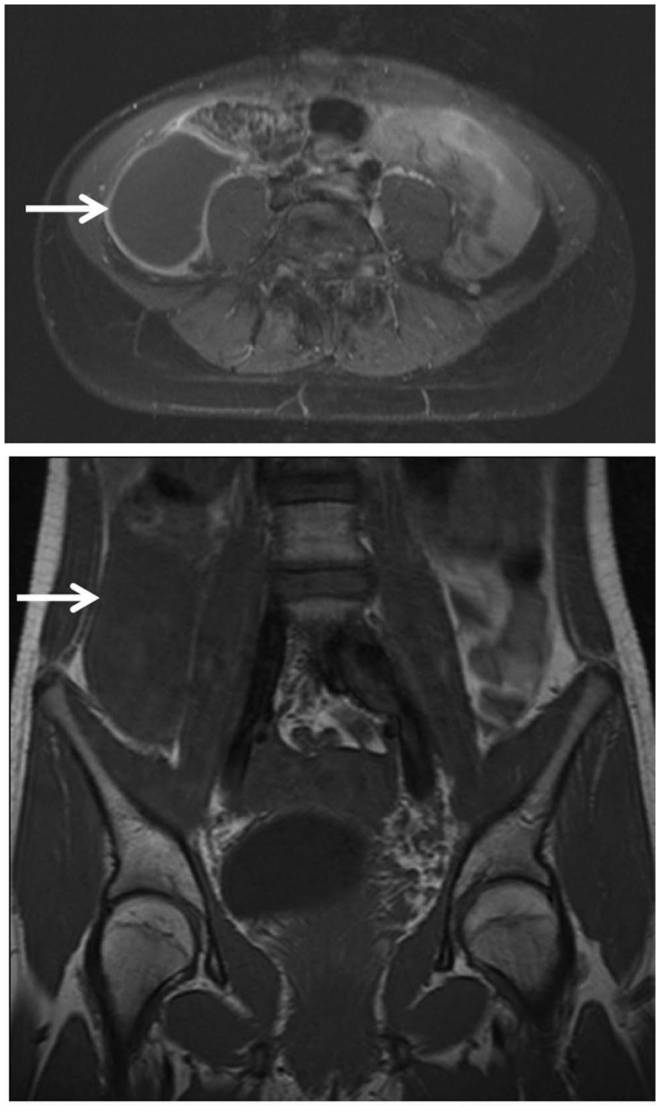

Figure 1. Magnetic resonance imaging demonstrating the right paracolic localization of the peritoneal inclusion cyst (arrow); axial view (top) and coronal view (bottom).

Dissection was commenced at the lower pole of the peritoneal inclusion cyst located lateral to the cecum **(Figure 2)**. The adhesions were dissected using the LigaSure (Covidien, Mansfield, MA) device **(Figure 3)**. No communication existed between the lumen of the colon and the cyst. However, due to the chronic inflammation of the peritoneal inclusion cyst, severe adhesions were encountered throughout and at times obscuring the plane of dissection **(Figure 4)**. When the cyst was dissected free from its attachments, a needle was introduced through the cyst wall to aspirate the cystic contents. An endoscopic specimen retrieval bag was used to remove the cyst wall. Care was also taken to identify the right ureter and to prevent

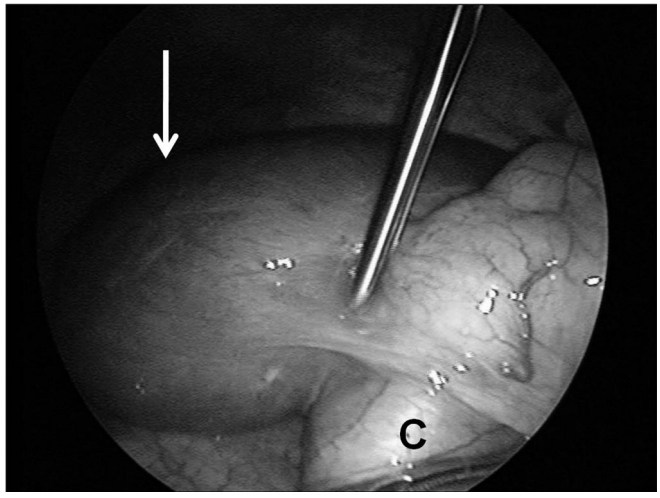

Figure 2. Laparoscopic view of the peritoneal inclusion cyst (arrow) approximated along the lateral border of the cecum (C).

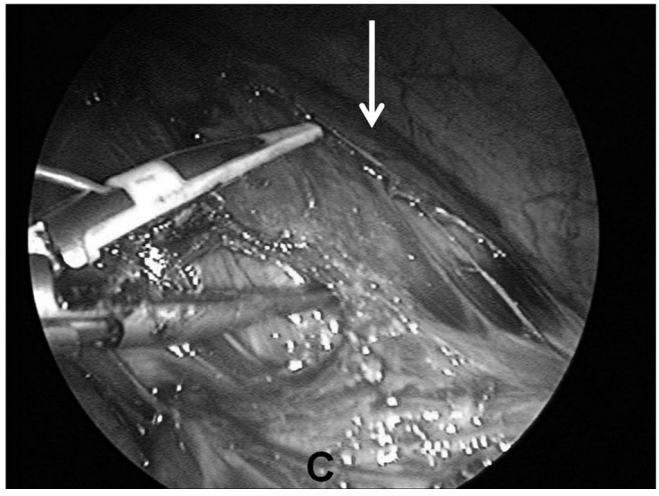

Figure 3. The LigaSure device was used to dissect between the plane of the peritoneal inclusion cyst (arrow) and the cecum (C).

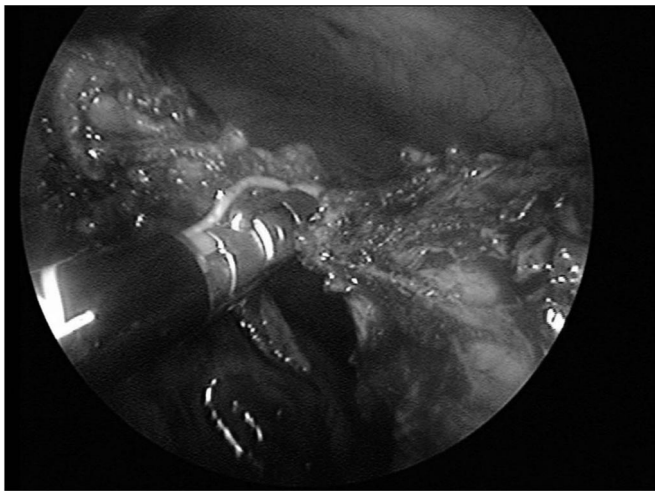

Figure 4. Severe adhesions were encountered between the peritoneal inclusion cyst and the colon closer to the hepatic flexure obscuring the plane of dissection.

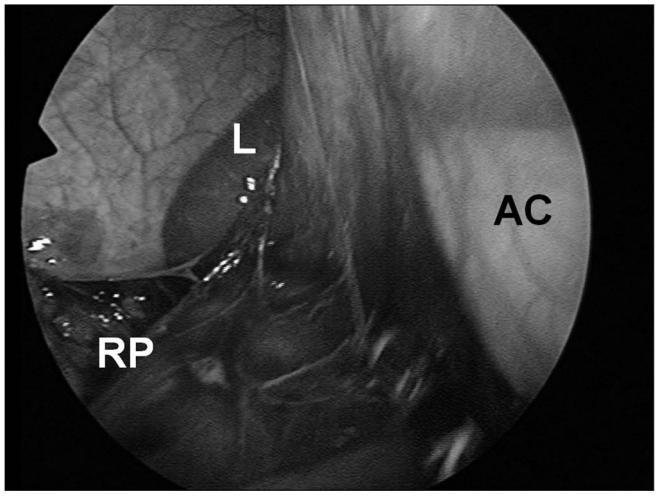

Figure 5. The retroperitoneal space (RP) exposed after the cyst resection. The dissection was performed up to the level of the liver (L)/Hepatic flexure along the lateral length of the ascending colon (AC).

injury during dissection between the cyst extended along the entire length of the ascending colon up to the hepatic flexure **(Figure 5)**. A concomitant elective appendectomy was also performed. The procedure was completed in 75 minutes. Histology demonstrated an epithelial fibrous cyst wall with a chronic recurrent phlegmonous inflammatory reaction.

DISCUSSION

Peritoneal inclusion cysts have not been observed or reported to date in prepubertal females. Although these cysts have been reported to occur in multiples,[3] the case presented herein demonstrates a solitary cyst with a paracolic extension. The paracolic extension of the cyst and its proximity to the lateral wall of the ascending colon led to the presumption of colon duplication in the differentials. It was also presumed that the inflammation of the duplicated colon was responsible for the acute abdominal pain. However, no luminal continuity between the cysts and the colon could be identified, and the reason for the infection of the cyst was not confirmed. Bacterial translocation from the colon can be hypothesized to be the cause of recurrent inflammation of the cyst.

The minimal access approach toward the management of peritoneal inclusion cysts has been reported in an adult female with removal and drainage of the mass.[5] Further reports with minimal access surgical options for these cysts do not exist for either the adult or the adolescent population. The laparoscopic approach enabled direct visualization of the cyst and complete dissection and removal with the advantages associated with minimal access surgery. The application of a tissue-sealing device was advantageous in this surgery.

Because the patient presented with known congenital lower extremity venous valve agenesis, minimal abdominal insufflation pressures were used to prevent venous stasis in the lower extremities. Congenital lower extremity venous valve agenesis with the absence of valves both in the superficial and deep veins is seldom reported in the literature, and it has been suggested that this may be because the condition is underdiagnosed.[6] This type of valve agenesis is genetic, because familial congenital absence of valves in the deep leg veins has been reported.[7] Although this condition has relevance for minimal access surgery with the consequences of prolonged venous stasis during prolonged procedures, the literature is void of reports of the association between laparoscopic surgery and the clinical condition of congenital lower extremity venous valve agenesis.

Critical review of laparoscopic procedures has shown its association with thromboembolic complications.[8] Thrombosis may be caused by detrimental effects of pneumoperitoneum on venous flow (increased abdominal pressure and negative Trendelenburg position) and activation of the hemostatic system. The effect of pneumoperitoneum on the venous flow of lower extremities has been further investigated and significant differences were found when pressures of 11mm Hg were compared with 14mm Hg with a significant increase in cross-sectional areas of

the femoral and saphenous vein when high pressures were used.[9]

Because our patient was a child, the lower pressure of 8mm Hg was used for the surgical procedure, because the risk factor of congenital lower extremity venous valve agenesis was known. However, if this condition is previously known in adolescents or adults, it may be beneficial to use intermittent sequential compression of the lower limbs to prevent venous stasis in laparoscopic surgery as suggested for colorectal procedures.[10] Although, there were no complications and our patient had a 2-year follow-up, the possible impact of thrombotic complications in patients with congenital lower extremity venous valve agenesis in the development of thrombosis after laparoscopic surgery must be carefully considered.

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